

000000000000

FIGURE 1A


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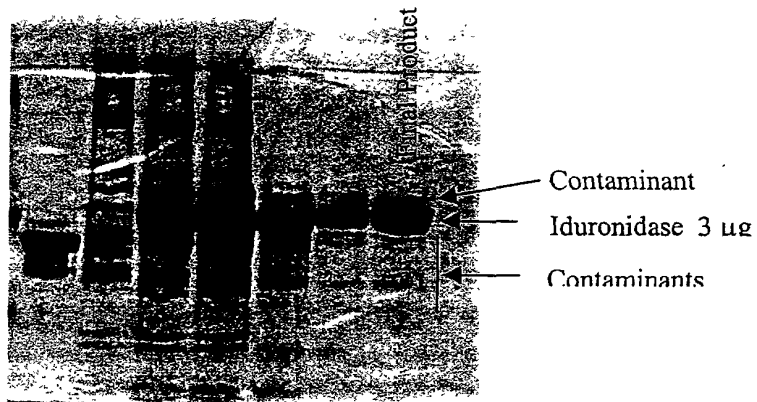
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      3760      3770      3780      3790      3800      3810      3820
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CGCTGATCAG CCTCGACTGT GCCTTCTAGT TGCCAGCCAT CTGTTGTTTG CCCCTCCCCC GTGCCTTCCT
      3830      3840      3850      3860      3870      3880      3890
      *      *      *      *      *      *      *
TGACCCTGGA AGGTGCCACT CCCACTGTCC TTCCCTAATA AAATGAGGAA ATTGCATCGC ATTGTCTGAG
      3900      3910      3920      3930      3940      3950      3960
      *      *      *      *      *      *      *
TAGGTGTCAT TCTATTCTGG GGGGTGGGGT GGGGCAGGAC AGCAAGGGGG AGGATTGGGA AGACAATAGC
      3970      3980      3990      4000      4010      4020      4030
      *      *      *      *      *      *      *
AGGCATGCTG GGGATGCGGT GGGCTCTATG GCTTCTGAGG CGGAAAGAAC CAGCTGGGGC TCGAGAGCTT
      4040      4050      4060      4070      4080      4090      4100
      *      *      *      *      *      *      *
GGCGTAATCA TGGTCATAGC TGTTCCTGT GTGAAATTGT TATCCGCTCA CAATTCCACA CAACATACGA
      4110      4120      4130      4140      4150      4160      4170
      *      *      *      *      *      *      *
GCCGGAAGCA TAAAGTGTAAGCCTGGGGT GCCTAATGAG TGAGCTAACT CACATTAATT GCGTTGCCCT
      4180      4190      4200      4210      4220      4230      4240
      *      *      *      *      *      *      *
CACTGCCCGC TTTCCAGTCG GGAAACCTGT CGTGCCAGCT GCATTAATGA ATCGGCCAAC GCGCGGGGAG
      4250      4260      4270      4280      4290      4300      4310
      *      *      *      *      *      *      *
AGGCGGTTTG CGTATTGGGC GCTCTTCCGC TTCTCGCTC ACTGACTCGC TCGCTCGGT CGTTCGGCTG
      4320      4330      4340      4350      4360      4370      4380
      *      *      *      *      *      *      *
CGGCGAGCGG TATCAGCTCA CTCAAAGGCG GTAATACGGT TATCCACAGA ATCAGGGGAT AACGCAGGAA
      4390      4400      4410      4420      4430      4440      4450
      *      *      *      *      *      *      *
AGAACATGTG AGCAAAAGGC CAGCAAAAGG CCAGGAACCG TAAAAAGGCC GCGTTGCTGG CGTTTTTCCA
      4460      4470      4480      4490      4500      4510      4520
      *      *      *      *      *      *      *
TAGGCTCCGC CCCCTGACG AGCATCACAA AAATCGACGC TCAAGTCAGA GGTGGCGAAA CCCGACAGGA
      4530      4540      4550      4560      4570      4580      4590
      *      *      *      *      *      *      *
CTATAAAGAT ACCAGGCGTT TCCCCCTGGA AGCTCCCTCG TCGCTCTCC TGTTCCGACC CTGCCGCTTA
      4600      4610      4620      4630      4640      4650      4660
      *      *      *      *      *      *      *
CCGGATACCT GTCCGCCTTT CTCCCTTCGG GAAGCGTGCC GCTTTCTCAA TGCTCAGCT GTAGGTATCT
      4670      4680      4690      4700      4710      4720      4730
      *      *      *      *      *      *      *
CAGTTCGGTG TAGGTCGTTT GCTCCAAGCT GGGCTGTGTG CACGAACCCC CCGTTCAGCC CGACCGCTGC
      4740      4750      4760      4770      4780      4790      4800
      *      *      *      *      *      *      *
GCCTTATCCG GTAACATCG TCTTGAGTCC AACCCGGTAA GACACGACTT ATCGCCACTG GCAGCAGCCA

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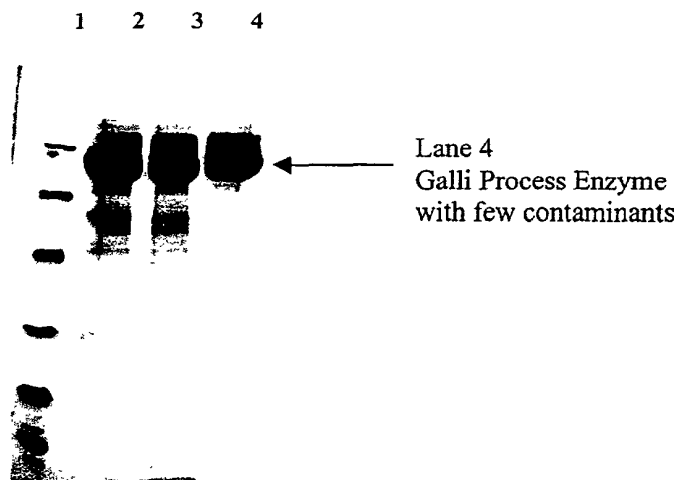
FIGURE 1E

FIGURE 2. SDS-POLYACRYLAMIDE GELS DEMONSTRATING IMPROVEMENTS IN PURITY

Gel using the Kakkis et al 1994, published procedure for purification



Gel using the new Galli Process contained in this application



1. Molecular Weight Marker
2. Prior Process Carson (nonpublished) Batch 2000C9001 Reference Reduced (7.5 μ g)
3. Same Batch 2000C9001 Reference Reduced (5.0 μ g)
4. Galli Process Enzyme Batch P10006 (5.0 μ g)

FIGURE 2

FIGURE 3A IDURONIDASE PRODUCTION USING THE GALLI PROCESS

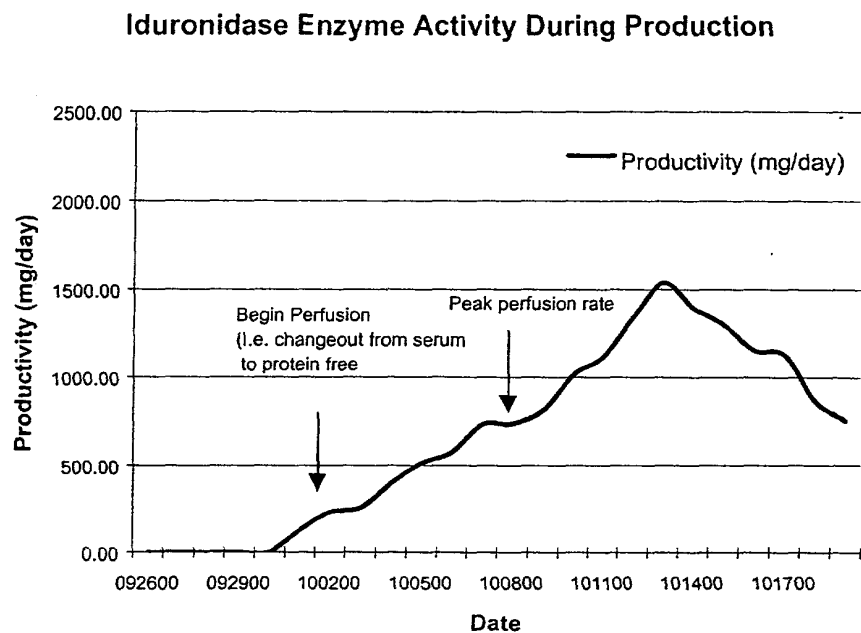
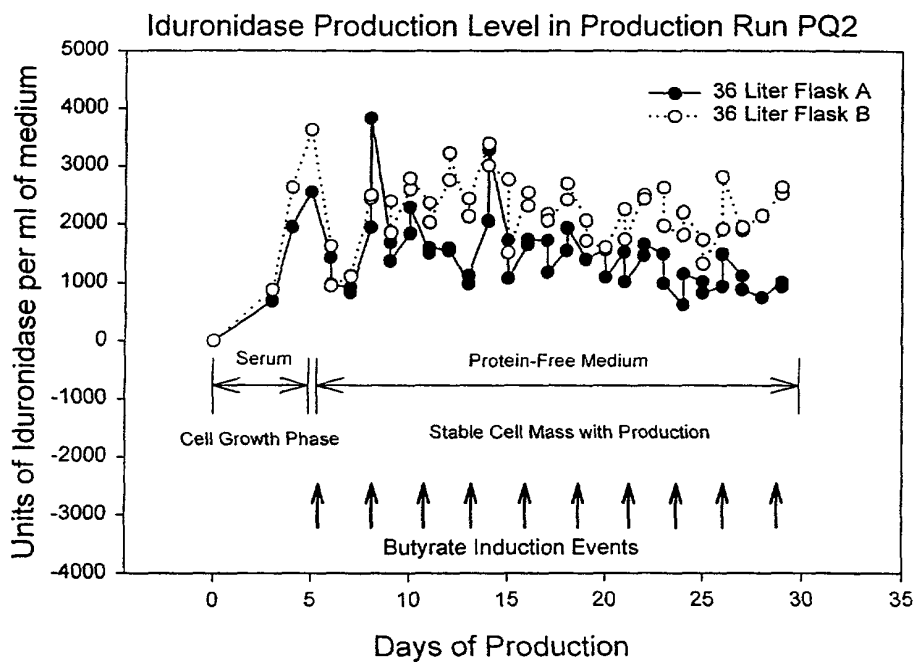


FIGURE 3B. IDURONIDASE PRODUCTION USING BUTYRATE INDUCTION



Reduction in Liver Volume During Enzyme Therapy

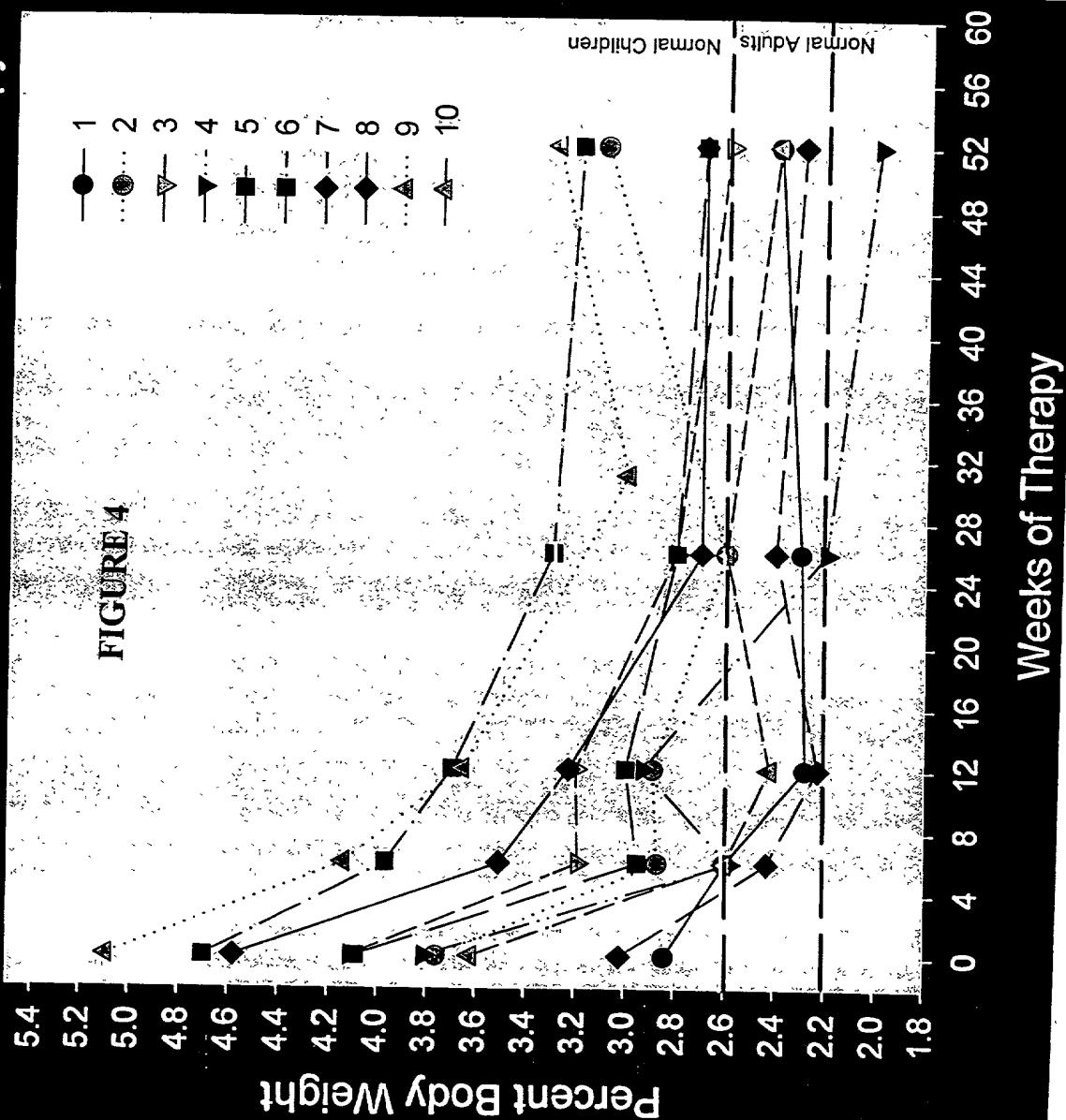


FIGURE 4

Urinary GAG Excretion During Enzyme Therapy

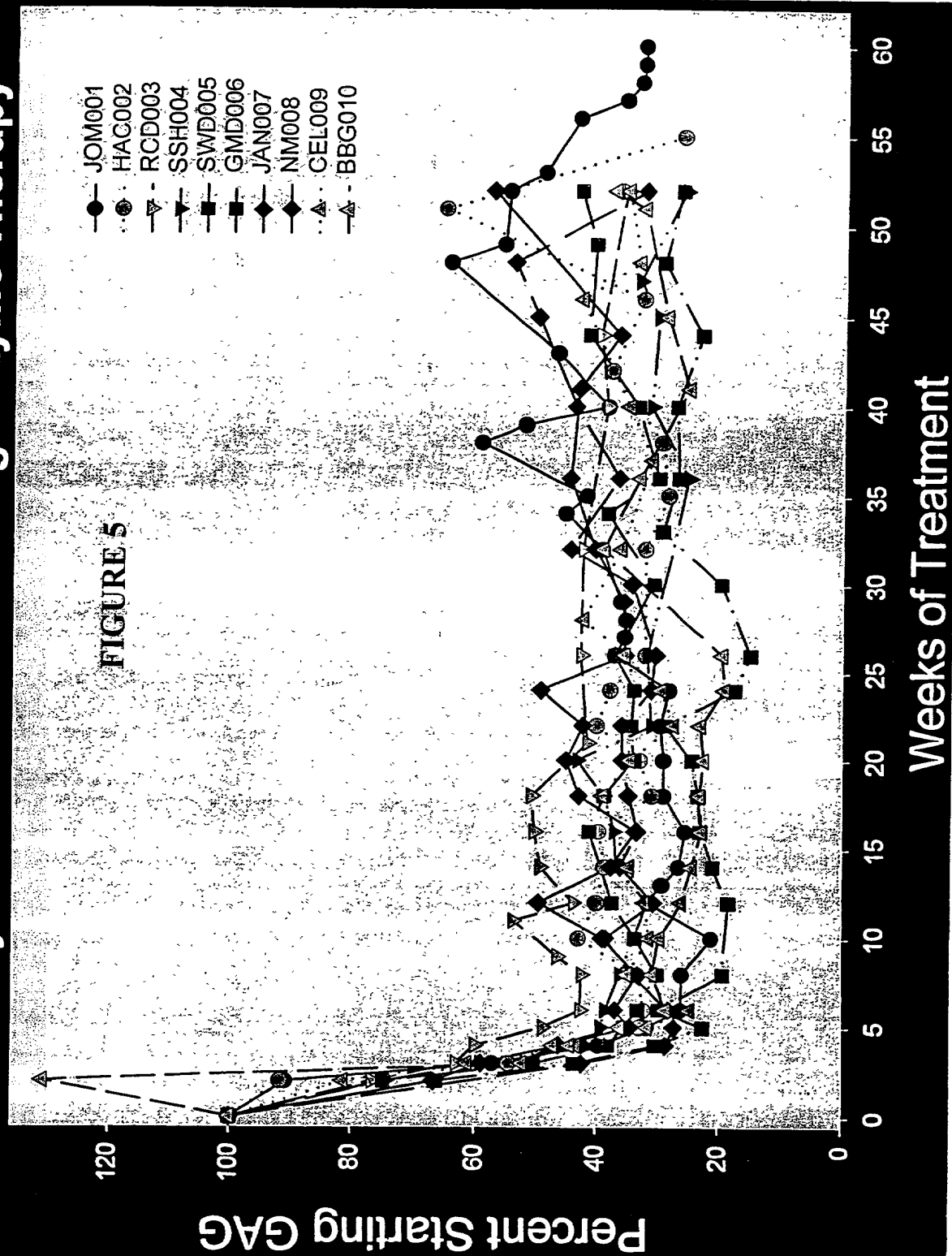


FIGURE 5

Elbow and Knee Extension in HAC002

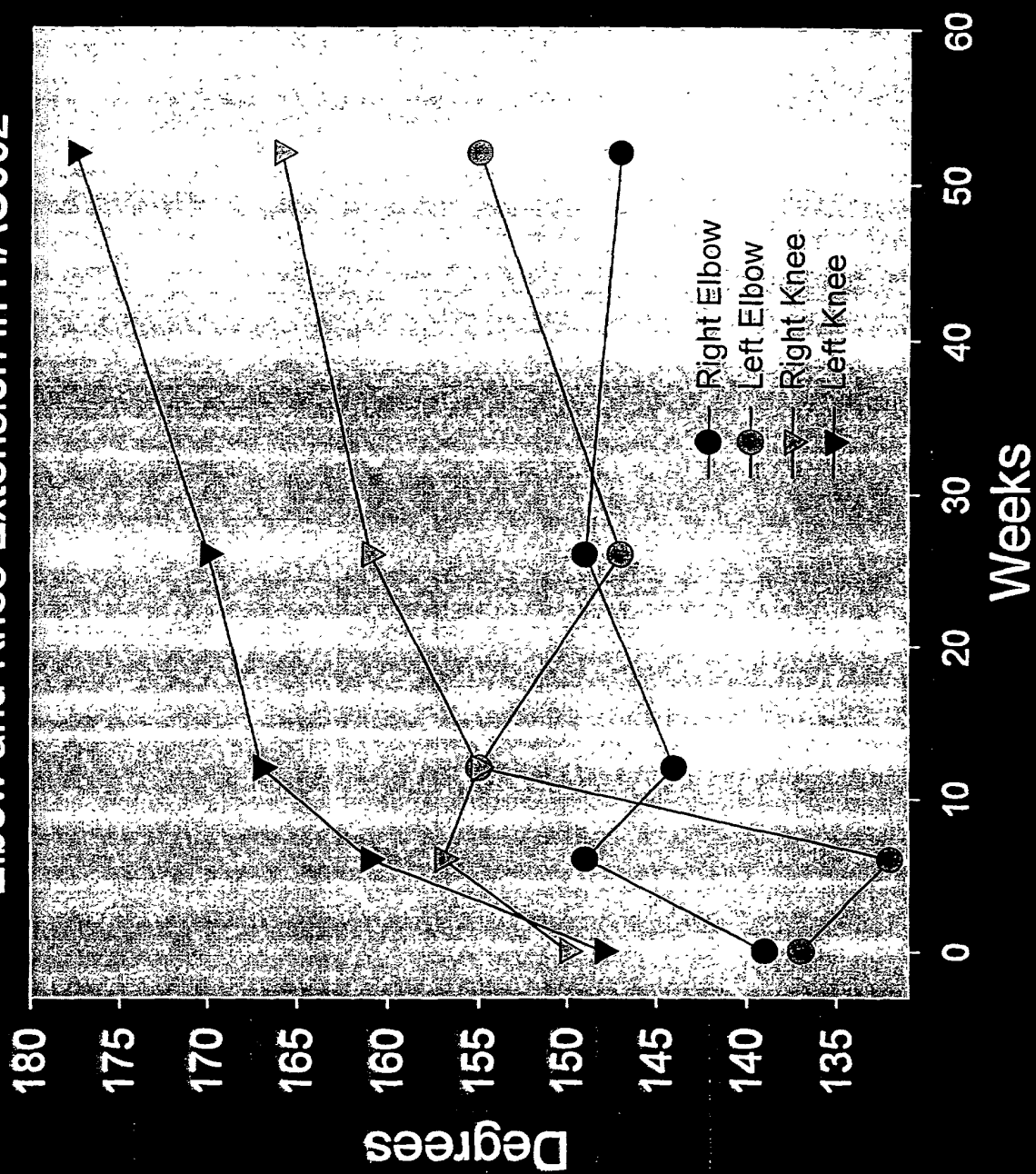


FIGURE 6

Shoulder flexion to 104 weeks in four patients with most restriction

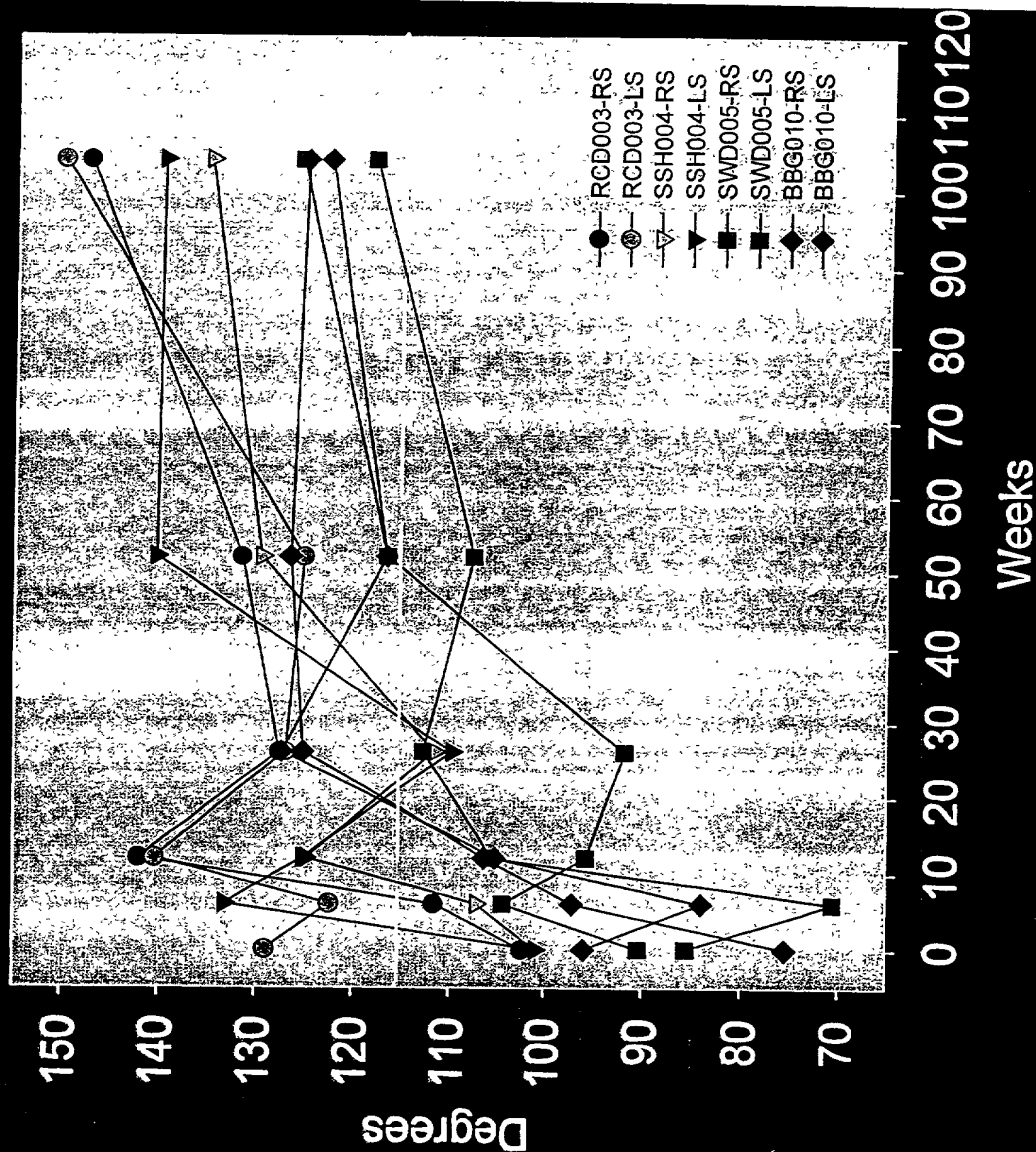
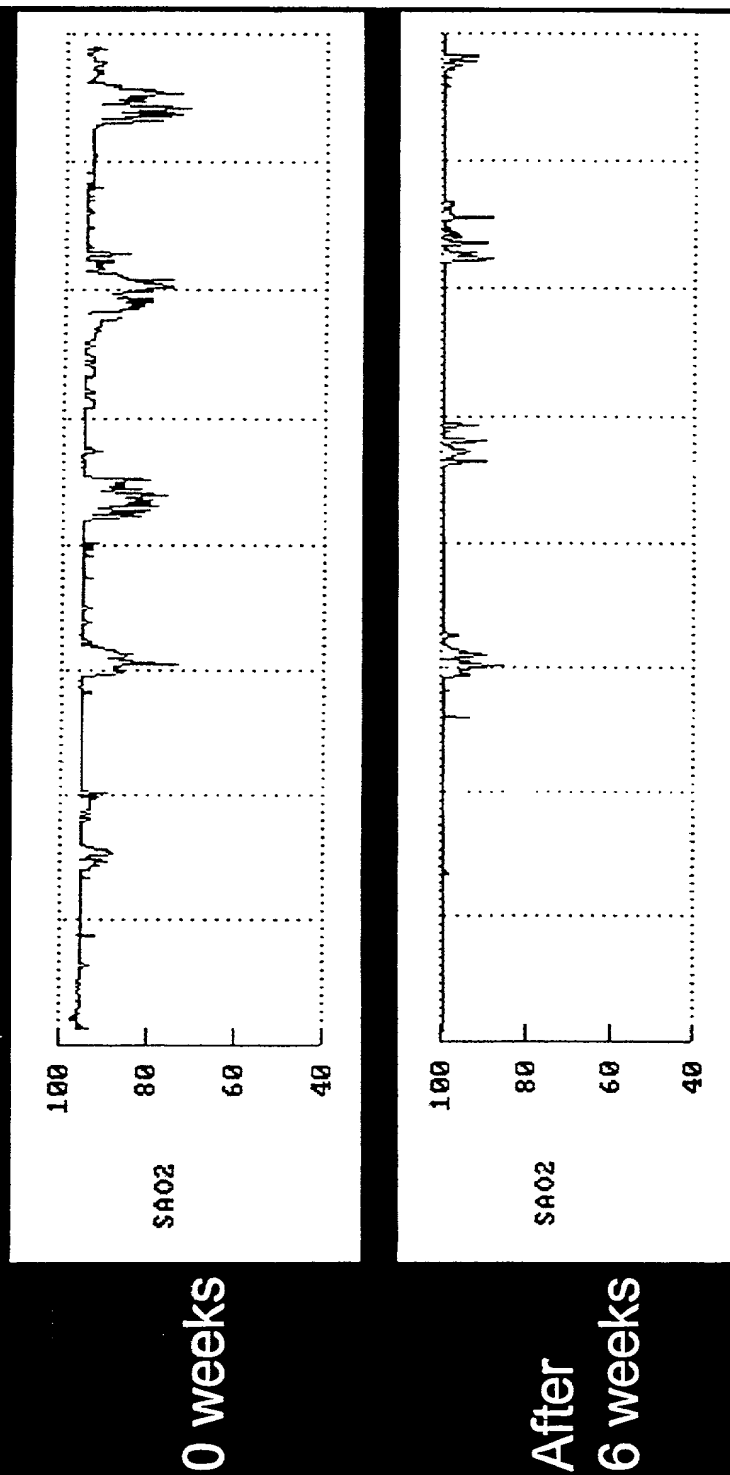


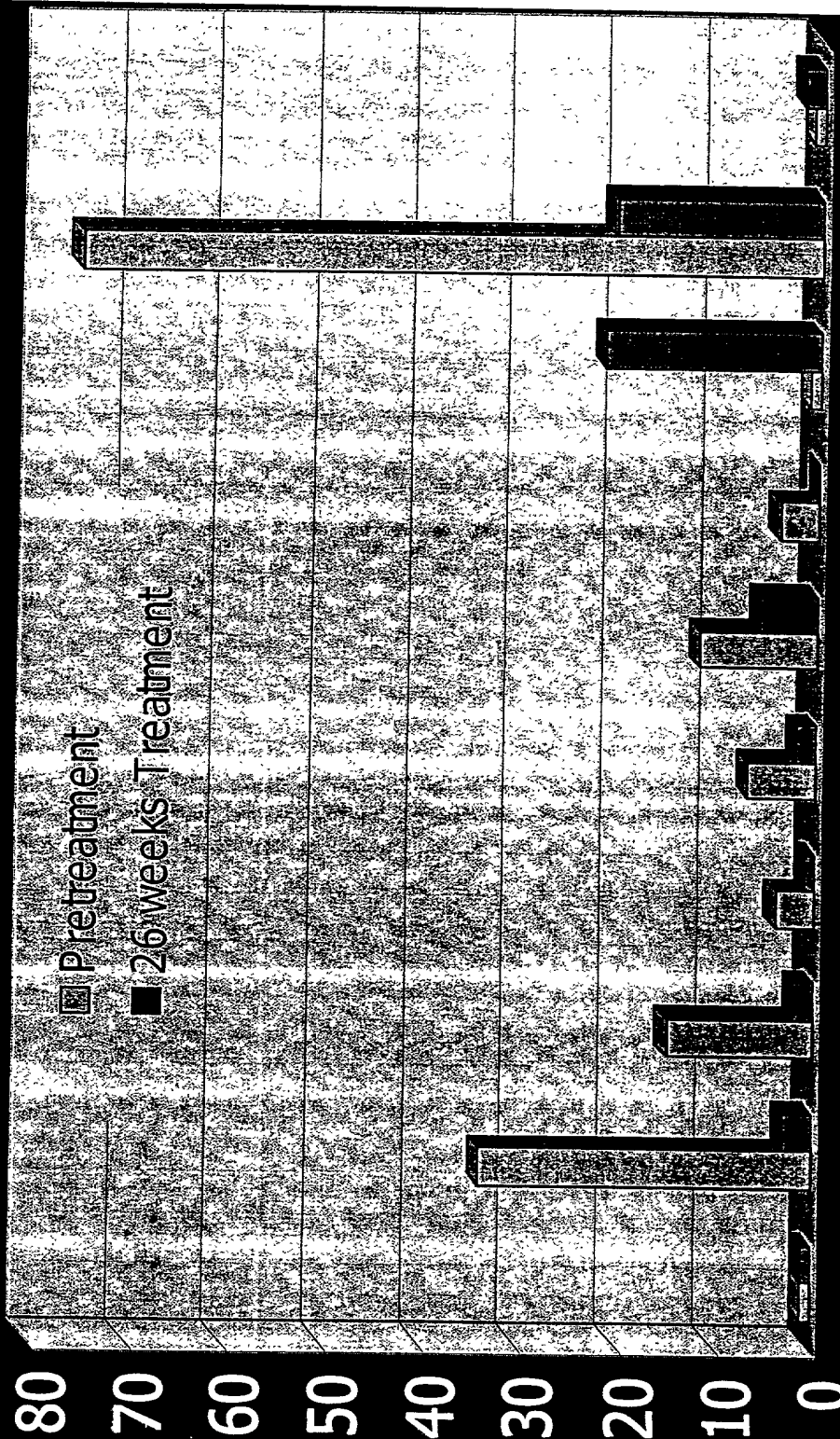
FIGURE 7

FIGURE 8

Sleep Apnea Improves



Apneas + Hypopneas During Sleep Pre and Post Treatment



JOM001 AHC002 RCD003 SSH004 SWD005 GMD006 JAN007 N-M008 CEL009 BBG010

FIGURE 9

Pulmonary Function Tests in GMD006

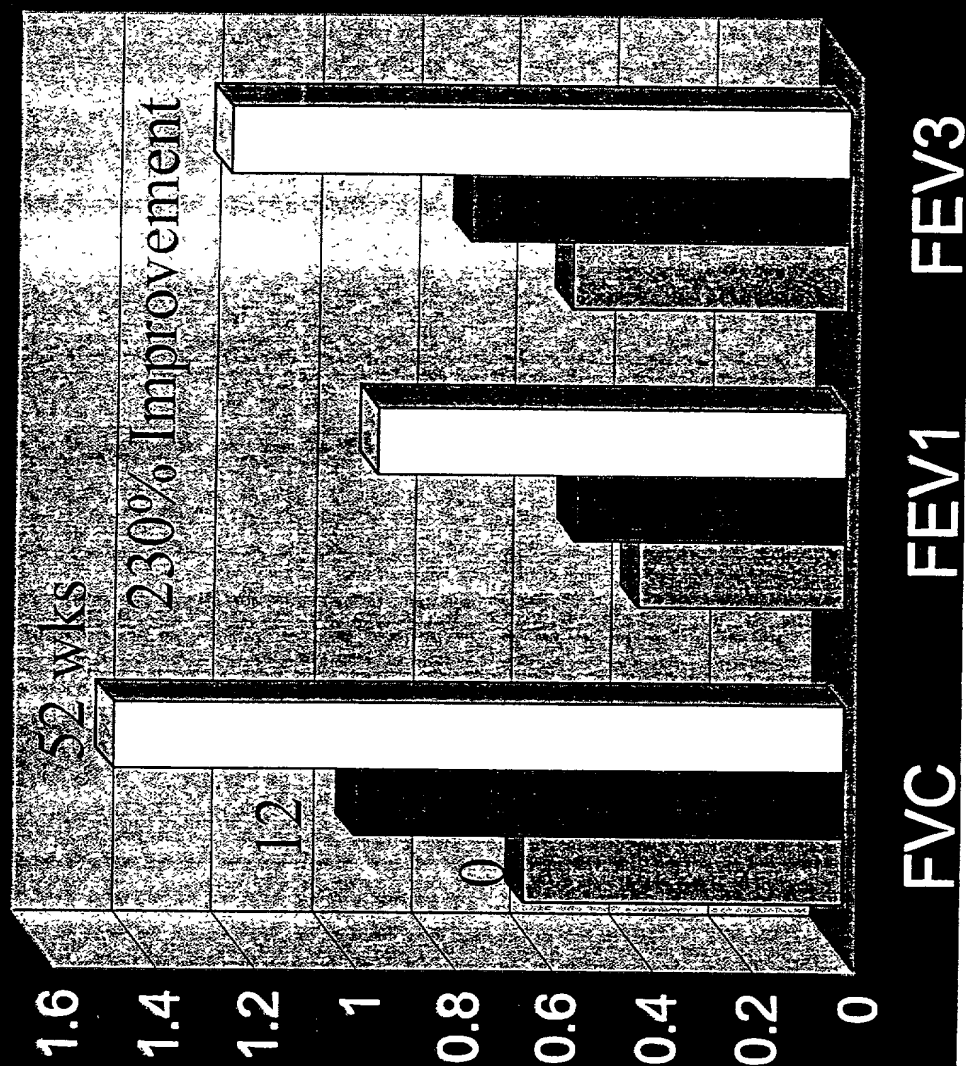


FIGURE 10

Increased Height Growth Velocity

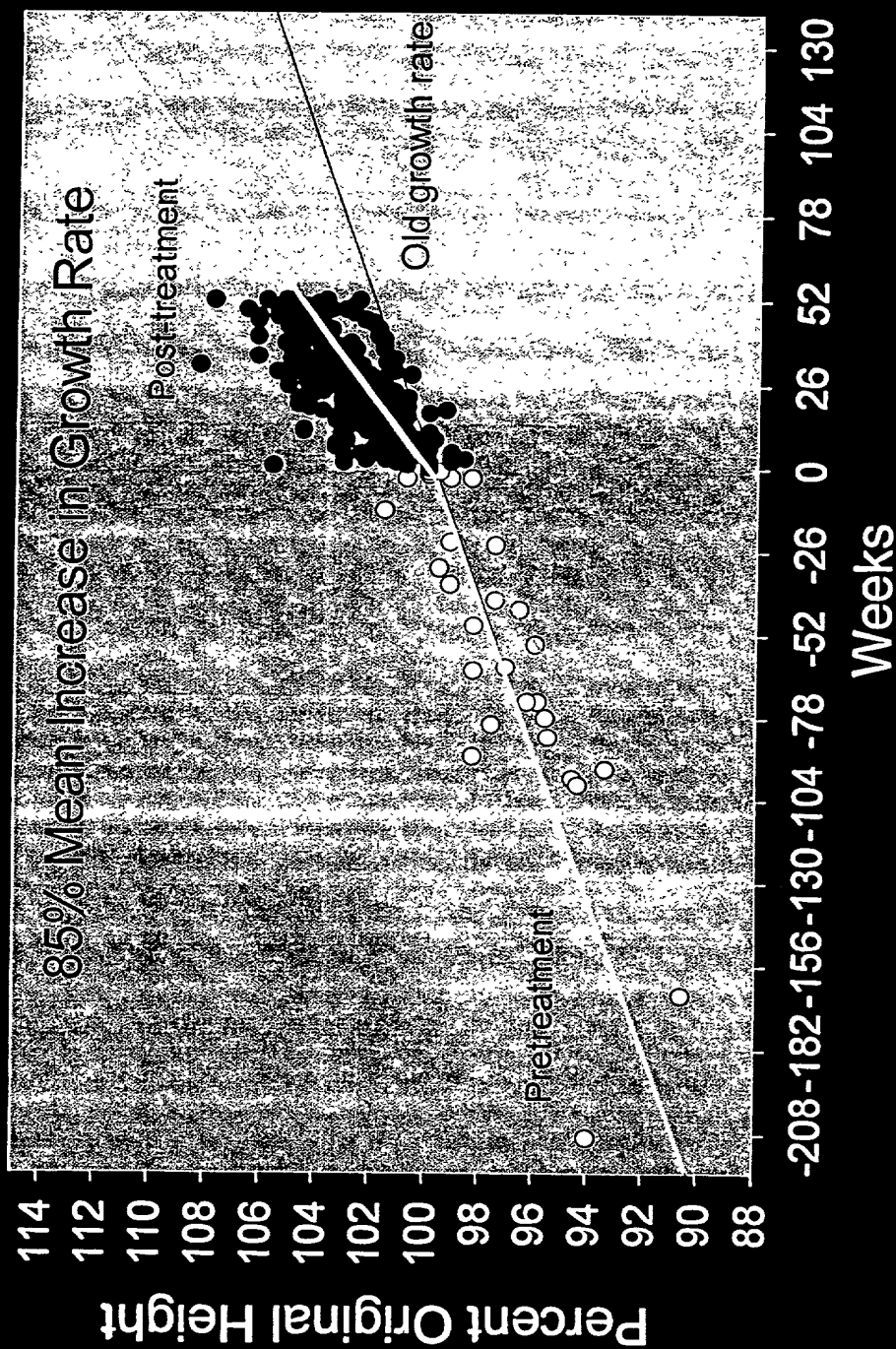


FIGURE 11

FIGURE 12.

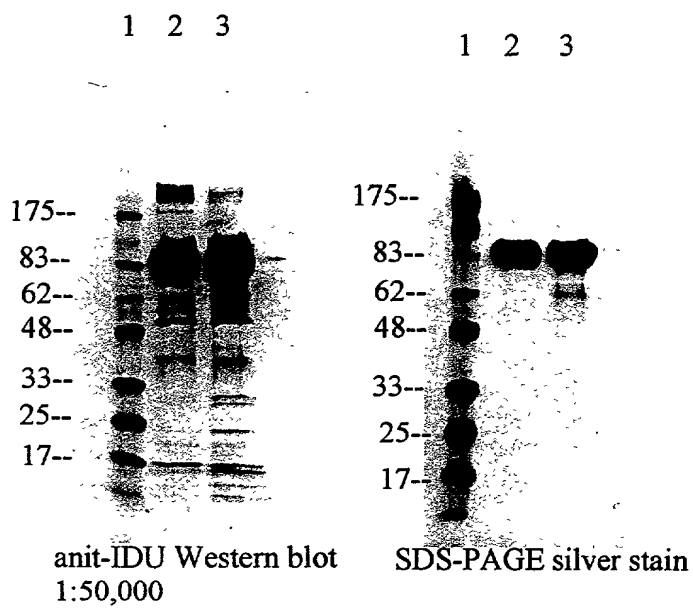
COMPARISON OF HOST PROTEIN CONTAMINATION BETWEEN A PRIOR AND THE NEW GALLI PROCESS

Chinese Hamster Ovary Host Protein Contamination by ELISA Assay

SOURCE AND BATCH NUMBER	CHOP PROTEIN CONTAMINATION (microgram per milligram)	PERCENT CHOP CONTAMINATION	PURITY OF THE ENZYME FROM CHOP
Prior Process (Carson/REI)			
C9002	14	1.4%	98.6%
C9003	24	2.4%	97.6%
C9004	16	1.6%	98.4%
New Process (Galli)			
P1003	<1.3	<0.13%	>99.9%
P1006	1.2	0.12%	99.9%
P1007	<0.6	<0.06%	>99.9%
P1008	<0.67	<0.067%	>99.9%

FIGURE 12

Comparison of Galli and Carson Material



- 1 Marker
- 2 Galli Referenced-0201
- 3 Carson C9002

5ug/lane

FIGURE 13